

Wilkie syndrome. Review of the literature

Síndrome de Wilkie. Revisión de la literatura

Jesús Guadalupe Benavides-Olivera,* Noel Alonso Márquez-Jurado,†
Víctor Alejandro Salgado-Yáñez‡

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ABSTRACT

A 48-year-old male presented at the time of admission with a seven-day history of oral intolerance, accompanied by nausea and vomiting of gastric contents on multiple occasions, progressive abdominal pain, and distension. Physical examination revealed that he was in pain, cachectic, with distension and generalized abdominal pain, tympanic on percussion. A simple phase abdominal CT scan was performed, showing excessive dilatation of the stomach without intestinal dilatation; the diagnostic approach was complemented with a contrast CT scan identifying aortomesenteric impingement. The patient received conservative treatment for 48 hours without improvement, so a laparoscopic duodenojejunal anastomosis procedure was performed. There is still debate about diagnosing and treating superior mesenteric artery impingement syndrome since symptoms do not always correlate well with abnormal anatomical findings in radiological studies and may not resolve completely after treatment.

RESUMEN

Masculino de 48 años que presenta al momento de su ingreso un cuadro de siete días de evolución con intolerancia a la vía oral, acompañado de náusea y vómito de contenido gástrico en múltiples ocasiones, dolor y distensión abdominal progresiva. A la exploración física con facies de dolor, caquético, distensión y dolor abdominal generalizado, timpanismo a la percusión. Se realiza tomografía abdominal en fase simple donde se observa dilatación excesiva del estómago sin dilatación intestinal, se complementa abordaje diagnóstico con tomografía contrastada identificando pinzamiento aortomesentérico. El paciente recibió tratamiento conservador por 48 horas sin mejoría, por lo que se lleva a cabo procedimiento duodeno-yeyuno anastomosis laparoscópica. En la actualidad aún existe controversia en torno al diagnóstico y tratamiento del síndrome de pinzamiento de la arteria mesentérica superior, ya que los síntomas no siempre se correlacionan bien con los hallazgos anatómicos anormales en los estudios radiológicos, y pueden no resolverse por completo después del tratamiento.

INTRODUCTION

The superior mesenteric artery syndrome is an unusual cause of upper intestinal obstruction, known by various names such as Wilkie syndrome, duodenal arteriomesenteric obstruction, and chronic duodenal ileus.¹

It is characterized by compression of the third portion of the duodenum due to the narrowing of the space between the superior mesenteric artery and the aorta, attributed to the loss of the mesenteric fat pad.^{2,3}

Symptoms do not always correlate with abnormal anatomic findings in radiological studies.³

The characteristics of at-risk patients are decreased acuity of the aortomesenteric angle due to weight loss (greater than 10 kg), leading to loss of the mesenteric fat pad.

The syndrome is commonly associated with severe and debilitating diseases such as neoplasms, malabsorption syndromes, acquired immune deficiency syndrome, trauma, and burns.⁴

* General Surgery Service.

† General Surgery Resident.

‡ Internal Medicine Service.

Christus Muguerza Hospital Del Parque. Chihuahua, Mexico.

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The syndrome is most often described after corrective spine surgery for scoliosis, which causes a reduction of the aortomesenteric angle; under this circumstance, it is called “cast syndrome”.⁵

Rarely it is associated with a short ligament of Treitz that suspends the duodenum in an abnormally cephalic position.⁴

On clinical evaluation, patients may present acutely (after surgery) or more frequently with progressive symptoms. Symptoms are consistent with proximal small bowel obstruction, postprandial epigastric pain, and early satiety, and those with more advanced obstruction may have nausea, vomiting of biliary contents, and weight loss.⁵

Symptoms may be relieved in some positions, such as left lateral decubitus, ventral decubitus, or kneeling. These positions eliminate the tension of the aortomesenteric angle.⁶

Physical examination findings are not specific but may include abdominal distention, epigastric pain, and peristaltic rumbling.

Laboratory tests may be normal, and liver function and blood chemistry tests do not show alterations; patients with severe vomiting may present electrolyte abnormalities characterized by mild hypokalemia and metabolic alkalosis.^{5,6}

Diagnosis is often delayed and can lead to cardiac conduction disturbances, gastric perforation, obstruction by a duodenal bezoar, gastric pneumatosis, and portal venous gas.⁶

Differential pathology includes other causes of upper intestinal obstruction such as diseases with altered duodenal motility, type 2 diabetes, systemic sclerosis, and intestinal pseudo-obstruction.⁷

Diagnostic studies require a high index of suspicion since symptoms may be nonspecific. Diagnostic imaging criteria include: 1) Duodenal obstruction with an abrupt cut in the third portion. 2) Aortomesenteric angle less than 25° (considered the most sensitive measure of the diagnosis). 3) Aortomesenteric distance less than 8 mm. 4) High fixation of the duodenum by the ligament of Treitz.⁸

The initial treatment goals are to relieve obstructive symptoms by gastrointestinal decompression with a nasogastric tube and water and electrolyte correction.⁹

Nutritional support is essential. Enteral nutrition is preferred, and if it cannot be achieved orally, endoscopic placement of a nanojoule tube is suggested; total parenteral nutrition may be necessary if enteral nutrition is not possible.⁶

Some surgical options are the Strong’s procedure, gastro-jejunal anastomosis, and duodenojejunal anastomosis.

Strong’s procedure consists of derotational surgery aimed at repositioning the third and fourth portions of the duodenum to the right of the superior mesenteric artery in the case of a short Treitz ligament.⁸

Each of the surgical approaches has advantages and disadvantages. Strong’s procedure maintains bowel integrity; however, failure occurs in 25% of patients. Gastro-jejunal anastomosis decompresses the stomach but does not relieve duodenal obstruction, leading to blind loop syndrome or peptic ulcer. It is generally accepted that duodenojejunal-anastomosis has superior results to the previous ones.^{9,10}

CLINICAL CASE

The patient was a 48-year-old male residing in a nursing home with a history of cerebellar ataxia of a three-month duration that conditions prostration and appendectomy with open technique ten years ago.

At the time of her admission, he had seven days of evolution with oral intolerance, accompanied by nausea and vomiting of gastric content on multiple occasions, abdominal pain, and postprandial distension.

On physical examination, he was found with pain facies, cachectic, abdominal distension, and pain on palpation in the epigastric region, tympanic on percussion, no evidence of peritoneal irritation, absence of peristalsis, mucous membranes, and integuments with suboptimal hydration, in addition to spastic extremities. Laboratory studies revealed leukocytosis and hypoalbuminemia.

An abdominal CT scan was performed in which gastric dilatation was identified with loss of gastric folds and decreased distance between the aorta and superior mesenteric artery, measuring 8.05 mm (*Figure 1*). The

sagittal section of the CT scan showed an acute aortomesenteric angle of 14.92° (Figure 2). Since his admission, he received management with gastric decompression through a nasogastric tube, water and electrolyte replacement, analgesics, and antibiotic therapy with a third-generation cephalosporin antibiotic; however, the obstruction persisted despite conservative management for 48 hours, so surgical management was proposed and accepted. A laparoscopic approach with five ports was performed; the transverse colon was mobilized, and the site of obstruction was seen as secondary to clamping of the superior

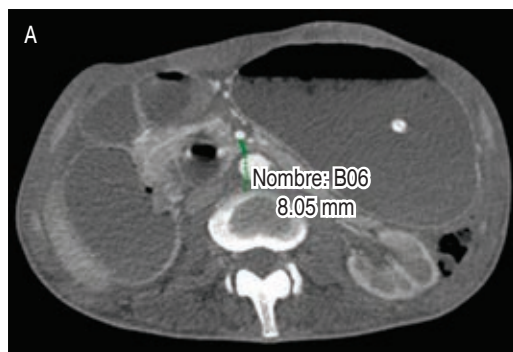


Figure 1: Contrast CT scan. **A)** Axial section showing a distance between the aorta and mesenteric artery of 8.05 mm. **B)** Coronal section showing gastric dilatation with loss of gastric folds and amputation of the third portion of the duodenum.



Figure 2: Contrast abdominal CT scan sagittal section, showing an acute aortomesenteric angle of 14.92° .

mesenteric artery between the third and fourth portion of the duodenum. A mechanical duodenojejunal-anastomosis was performed at 60 cm from the ligament of Treitz; the gastroenterostomy was closed in two planes with slow absorption monofilament suture for the first plane and non-absorbable for the second one. Postoperative evolution was good. He started drinking liquids at 24 hours and progressed to a soft diet at 48 hours. He was discharged home due to improvement 48 hours after the procedure. There have not been any subsequent clinical sequelae after two months. He has had adequate tolerance to the oral route, but the patient has not attended to control for post-surgical follow-up.

CONCLUSION

Willkie syndrome is a rare clinical entity whose diagnostic and therapeutic approach represents a clinical challenge. It is essential

to have a high clinical suspicion that allows identifying data of high intestinal obstruction and complementing it with laboratory and imaging studies that support the diagnosis and establish the most appropriate therapeutic approach for each case.

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Correspondence:

Víctor Alejandro Salgado-Yáñez, MD

E-mail: drvsalgado@gmail.com